PART XX The Cardiovascular System

حلول نيلسون mcq

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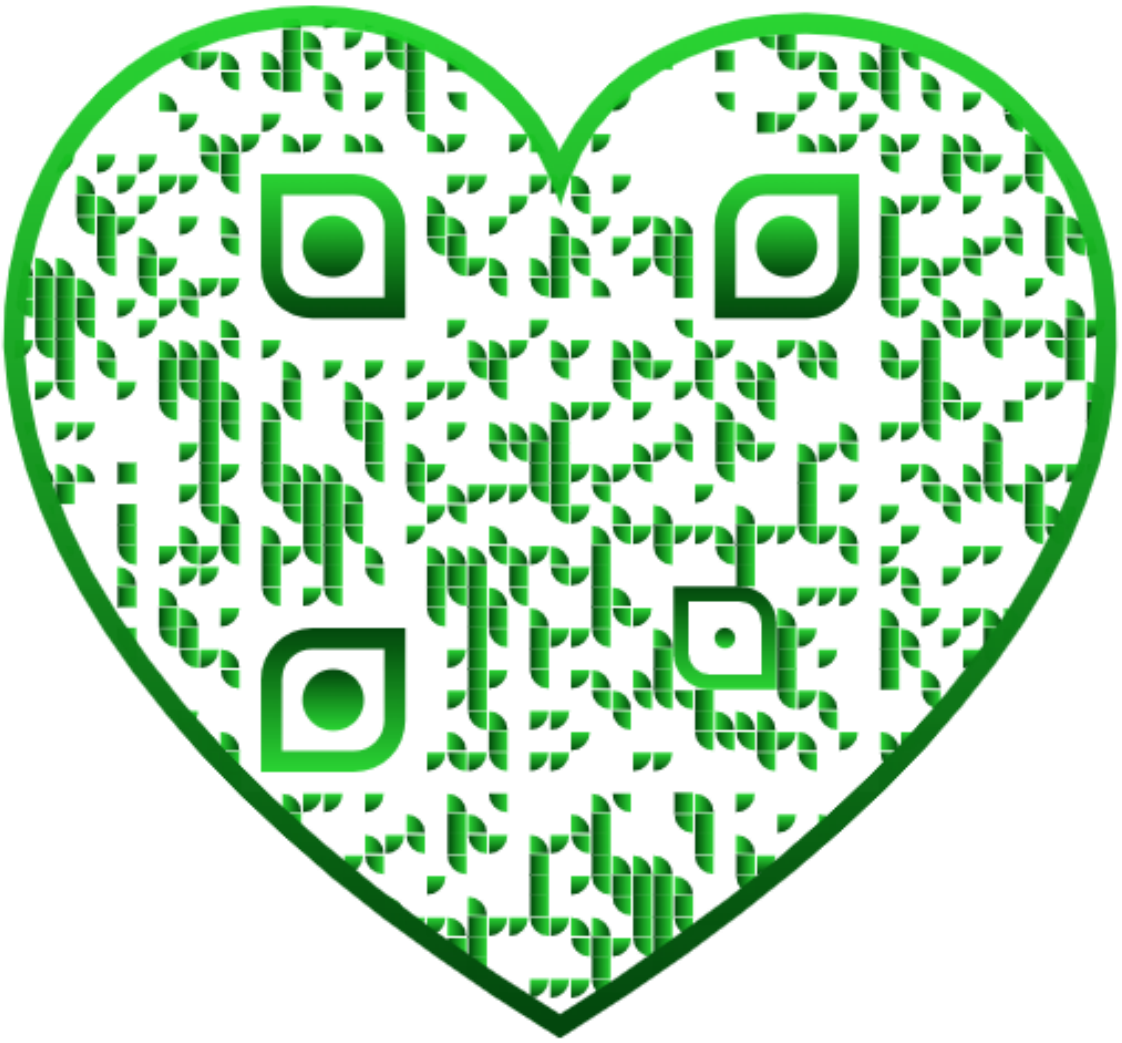
1. CHARGE association is characterized by the following cardiac feature
   A. endocardial cushion defect
   B. supravalvular aortic stenosis
   C. tetralogy of Fallot
   D. patent ductus arteriosus
   E. anomalous pulmonary venous return

2. Hypertrophic cardiomyopathy is a recognized association with
   A. infant of diabetic mother
   B. Marfan syndrome
   C. William syndrome
   D. trisomy 21 (Down syndrome)
   E. XO (Turner syndrome)

3. The following are characters of venous hum **EXCEPT**
   A. continuous murmur
   B. heard in infraclavicular region
   C. grades I-III/VI
   D. louder with patient in supine position
   E. changes with compression of jugular vein

4. Chest pain in the pediatric patient often generates a significant amount of patient and parental concern.
Of the following, the **MOST** common cause is
   A. myocarditis
   B. pericarditis
   C. pleurisy
   D. pneumothorax
   E. pneumonia

5. Congenital heart disease causing cyanosis without respiratory distress include the following **EXCEPT**
   A. tricuspid atresia
   B. aortic stenosis
   C. Ebstein anomaly
   D. pulmonary atresia



E. tetralogy of Fallot

6. The **MOST** common cyanotic cardiac lesion to present in the newborn period is
A. dextroposed transposition of the great arteries
B. hypoplastic left heart syndrome
C. tetralogy of Fallot
D. truncus arteriosus
E. pulmonary atresia

7. The ECG shows left ventricular hypertrophy and a superior QRS axis (between 0° and –90°) in
A. tricuspid atresia
B. Ebstein anomaly
C. pulmonary atresia
D. pulmonary stenosis
E. tetralogy of Fallot

8. The **MOST** common cause of death from cardiac defects in the first month of life is
A. d-Transposition without associated lesions
B. hypoplastic left heart syndrome
C. pulmonary atresia
D. truncus arteriosus
E. complex single ventricle

9. All the following are causes of heart failure in full-term neonate **EXCEPT**
A. asphyxial cardiomyopathy
B. coarctation of aorta
C. hypoplastic left heart
D. transposition of great arteries
E. ventricular septal defect

10. One of the following is a sign of right-sided heart failure
A. edema
B. tachypnea
C. orthopnea
D. wheezing
E. pulmonary edema

11. The **MOST** common cause of syncope in children is
A. Wolff-Parkinson-White syndrome
B. prolonged QT syndrome
C. atrioventricular block

D. neurocardiogenic syncope
E. myocarditis

12. Of the following, the **MOST** common clinical sign of coarctation of the aorta in older children is
A. cardiac enlargement
B. notching of the inferior border of the ribs
C. a systolic ejection click or thrill in the suprasternal notch
D. differential blood pressure: arms > legs
E. diminished or absent femoral or lower-extremity pulses

13. The chest x-ray may help to differentiate the types of congenital heart defects. Increased pulmonary markings (increased pulmonary blood flow) is seen in
A. pulmonary atresia
B. truncus arteriosus
C. tetralogy of Fallot
D. tricuspid atresia
E. Ebstein anomaly

14. The following are causes of congestive heart failure and cardiomegaly during the newborn period but no murmur **EXCEPT**
A. endocardial fibroelastosis
B. asphyxia
C. glycogen storage disease (Pompe disease)
D. paroxysmal supraventricular tachycardia
E. sepsis

15. SVT in children differ from physiologic sinus tachycardia by all the following **EXCEPT**
A. sudden onset and termination
B. persistent ventricular rate of >180 bpm
C. fixed RR interval on ECG
D. abnormal P-wave shape or axis or absent P waves
E. evident change in heart rate with activity

16. The following cardiac lesions are at increased risk for bacterial endocarditis **EXCEPT**
A. mitral insufficiency
B. aortic stenosis
C. atrial septal defect seccundum
D. coarctation of the aorta
E. patent ductus arteriosus

17. The following factors are most strongly associated with the development of coronary artery disease in patients with Kawasaki disease **EXCEPT**

A. duration of fever of >16 days
B. first-degree heart block
C. cardiomegaly
D. male gender
E. age of <1 year

18. The following are major side effects of PGE_1 **EXCEPT**

A. apnea
B. fever
C. cutaneous flushing
D. seizures
E. hypertension

19. One of the following cardiac lesions have been identified more in male

A. transposition of the great arteries
B. atrial septal defect
C. VSD
D. PDA
E. pulmonic stenosis

20. In **MOST** patients with an ASD, the characteristic physical finding is

A. a right ventricular systolic lift
B. a fixed splitted 2nd heart sound
C. a systolic ejection murmur
D. a short, rumbling mid-diastolic murmur
E. a mild left precordial bulge

21. Indications for surgical closure of a VSD include the following **EXCEPT**

A. patients in whom clinical symptoms and failure to thrive cannot be controlled medically
B. infants between 6 and 12 mo of age with large defects associated with pulmonary hypertension
C. patients older than 24 mo with a Qp : Qs ratio greater than 2 : 1.
D. patients with a supracristal VSD of any size
E. severe pulmonary vascular disease nonresponsive to pulmonary vasodilators

22. Regarding supracristal ventricular septal defect, the following are true **EXCEPT**

A. a supracristal VSD is complicated by aortic insufficiency in 50-90% of patients
B. the incidence is higher in Asian children

C. aortic insufficiency is most often not recognized until late in the 1st decade of life
D. the murmur of a supracristal VSD is usually heard at the mid to upper left sternal border
E. closure of supracristal ventricular VSDs at the time of diagnosis is not recommended in an asymptomatic child

23. All the following are signs of coarctation of the aorta **EXCEPT**
A. femoral pulses are weak or absent
B. bounding pulses of the arms
C. femoral pulse occurs slightly before the radial pulse
D. blood pressure in the legs is lower than that in the arms
E. precordial impulse and heart sounds are usually normal

24. All the following are causes of chronic pulmonary venous hypertension **EXCEPT**
A. congenital mitral stenosis
B. total anomalous pulmonary venous return with obstruction
C. peripheral pulmonary stenosis
D. left atrial myxomas
E. cor triatriatum

25. Regarding mitral valve prolapse; all the following are true **EXCEPT**
A. it is predominantly congenital
B. it is usually sporadic
C. it is more common in girls
D. the dominant abnormal signs are auscultatory
E. antibiotic prophylaxis is recommended during surgery and dental procedures

26. Sinus bradycardia is due to slow discharge of impulses from the sinus node, the heart's natural pacemaker. Sinus bradycardia in neonates is considered if sinus rate is
A. <100 beats/min
B. <90 beats/min
C. <80 beats/min
D. <70 beats/min
E. <60 beats/min

27. The following procedures may abort an attack of SVT **EXCEPT**
A. placing of the face in ice water
B. straining
C. breath holding
D. standing on head
E. ocular pressure

28. The following drugs may cause long Q-T syndromes (LQTS) **EXCEPT**
    A. trimethoprim/sulfamethoxazole
    B. erythromycin
    C. imipramine
    D. pentostam
    E. risperidone

29. The **MOST** common cause of death in competitive athletes is
    A. aortic stenosis
    B. hypertrophic cardiomyopathy
    C. coronary artery abnormalities
    D. myocarditis
    E. long Q-T syndrome (LQTS)

30. The leading causative agents for endocarditis in pediatric patients are
    A. group D enterococci
    B. viridans-type streptococci
    C. Pseudomonas aeruginosa
    D. fungal organisms
    E. Serratia marcescens

31. The following is a major Duke criterion for the diagnosis of endocarditis
    A. new valve regurgitant flow by echocardiography
    B. Osler nodes
    C. single positive blood culture
    D. serologic evidence of infection
    E. high erythrocyte sedimentation rate

32. The lower limit of pulse rate in neonate at rest is
    A. 50/min
    B. 60/min
    C. 70/min
    D. 80/min
    E. 90/min

33. Careful evaluation of the character of the pulses is an important early step in the physical diagnosis of congenital heart disease. A wide pulse pressure with bounding pulses may suggest the following **EXCEPT**
    A. patent ductus arteriosus
    B. aortic insufficiency
    C. cardiomyopathy
    D. anemia

E. anxiety

34. Tall (>2.5 mm), narrow, and spiked P waves are seen in
A. Ebstein anomaly
B. ventricular septal defect [VSD]
C. patent ductus arteriosus [PDA]
D. severe mitral stenosis
E. mitral regurgitation

35. In utero heart failure, often with fetal pleural and pericardial effusions and generalized ascites (non-immune hydrops fetalis) may occur in
A. ventricular septal defect
B. coarctation of aorta
C. d-Transposition of great arteries
D. Ebstein anomaly
E. single ventricle

36. Of the following, the cardiac lesion resulting in increased volume load is
A. valvular pulmonic stenosis
B. patent ductus arteriosus
C. valvular aortic stenosis
D. coarctation of the aorta
E. mitral stenosis

37. Of the following, the cyanotic cardiac lesion with decreased pulmonary blood flow is
A. transposition of the great vessels
B. single ventricle
C. truncus arteriosus
D. tricuspid atresia
E. total anomalous pulmonary venous return without obstruction

38. The predominant manifestations of primary pulmonary hypertension include the following **EXCEPT**
A. exercise intolerance
B. precordial chest pain
C. syncope
D. low arterial oxygen saturation
E. dizziness

39. The following are common side effects of sildenafil **EXCEPT**
A. flushing

B. elevated liver function tests
C. headache
D. diarrhea
E. myalgia

40. Postoperative (cardiac surgery) pulmonary hypertension can be managed with
A. hyperventilation and inhaled nitric oxide
B. catecholamines
C. phosphodiesterase inhibitors
D. nitroprusside
E. diuretics

41. Patients who have undergone surgery entailing the use of cardiopulmonary bypass, should be watched carefully for
A. epilepsy
B. learning disabilities
C. anemia
D. pulmonary hypertension
E. arrhythmias

42. The postpericardiotomy syndrome is characterized by all the following **EXCEPT**
A. fever
B. decreased appetite
C. listlessness
D. vomiting
E. chest pain

43. For one of the following, total repair really achieved, with no requirement for long-term follow-up
A. atrial septal defects
B. ventricular septal defects
C. valvar pulmonic stenosis
D. uncomplicated tetralogy of Fallot
E. isolated patent ductus arteriosis

44. Pregnancy is contraindicated in mothers with
A. severe pulmonary hypertension
B. ventricular septal defects
C. prosthetic valves
D. atrial septal defects
E. unoperated cyanotic congenital heart disease

45. Surgical repair of VSD at the time of diagnosis should be considered in
    A. small supracristal VSD
    B. nonrestrictive VSD
    C. hemodynamically significant VSD
    D. restrictive VSD
    E. VSD with heart failure

46. The **MOST** common long-term complication for those patients who underwent early repair of complete atrioventricular canal before the development of pulmonary vascular disease is
    A. left AV valve regurgitation
    B. subaortic stenosis
    C. residual ventricular level shunts
    D. complete heart block
    E. endocarditis

47. Patients with a small PDA have the following criteria **EXCEPT**
    A. continuous murmur heard best at the left upper sternal border
    B. normal peripheral pulses
    C. normal pulmonary artery pressure by echocardiography
    D. asmptomatic and live a normal life expectancy
    E. risk for endocarditis is extremely low

48. Severe pulmonary valve stenosis defined as a peak gradient of
    A. >40 mm Hg
    B. >45 mm Hg
    C. >50 mm Hg
    D. >55 mm Hg
    E. >60 mm Hg

49. Symptoms in patients with aortic stenosis depend on the severity of the obstruction. Critical aortic stenosisis associated with the following **EXCEPT**
    A. cardiomegaly
    B. pulmonary edema
    C. weak pulses in all extremities
    D. pale skin
    E. maximum intensity of the murmur

50. All the following are true regarding the diagnosis of coarctation of the aorta **EXCEPT**
    A. cardiac enlargement are noted with severe coarctation
    B. notching of the ribs is common by late childhood
    C. doppler is useful for demonstrating the specific site of the obstruction

D. CT and MRI are valuable noninvasive tools for evaluation of coarctation when the echocardiogram is equivocal
E. diagnostic catheterization is mandatory before surgery

51. Shone complex consist of the following **EXCEPT**
A. coarctation of the aorta
B. subvalvar aortic stenosis
C. valvar aortic stenosis
D. pulmonic stenosis
E. mitral stenosis

52. Paroxysmal hypercyanotic attacks (hypoxic, "blue," or "tet" spells) are a particular problem during the 1st 2 yr of life. They are characterized by
A. early evening occurrence
B. an increase in intensity of the systolic murmur
C. unpredictable onset
D. metabolic alkalosis
E. more freguent spells in patients with marked cyanosis at rest

53. Depending on the frequency and severity of hypercyanotic attacks in tetralogy of Fallot, all the following procedures are true **EXCEPT**
A. placement of the infant in the knee-chest position
B. administration of oxygen
C. injection of morphine subcutaneously
D. rapid correction with intravenous sodium bicarbonate irrespective of the spell severity
E. intravenous administration of propranolol

54. The typical radiologic configuration in tetralogy of Fallot as seen in the anteroposterior view consists of all the following **EXCEPT**
A. normal heart size
B. some elevation of the cardiac apex
C. convexity in the region of the main pulmonary artery
D. right-sided aortic arch
E. diminished pulmonary vascularity

55. The severity of symptoms of Ebstein anomaly and the degree of cyanosis are highly variable and depend on the extent of
A. cardiac dysrhythmias
B. displacement of the tricuspid valve
C. atrial right-to-left shunt
D. pulmonary vascular resistance

E. polycythemia

56. D-transposition of the great vessels, a common cyanotic congenital anomaly, accounts for ≈5% of all congenital heart disease. In this anomaly, all the following are true **EXCEPT**

A. the systemic veins return to the right atrium
B. the pulmonary veins return to the left atrium
C. the connections between the atria and ventricles are normal
D. the aorta is posterior and to the right of the pulmonary artery
E. the aorta is anterior and to the right of the pulmonary artery

57. Obstructed total anomalous pulmonary venous return (TAPVR) is a pediatric cardiac surgical emergency because

A. prostaglandin therapy is usually not effective
B. severe pulmonary congestion develop
C. left atrium may be small
D. left ventricle may be small
E. pulmonary hypertension develop

58. In neonates with total anomalous pulmonary venous return and marked pulmonary venous obstruction, the chest x-ray demonstrates

A. small heart
B. enlarged heart
C. prominent pulmonary artery
D. prominent right ventricle
E. subaortic stenosis

59. In truncus arteriosus, a single arterial trunk arises from the heart and supplies the systemic, pulmonary, and coronary circulations. All the following are true **EXCEPT**

A. VSD is always present
B. the truncus overriding the defect
C. both ventricles are at systemic pressure
D. heart failure ensues immediately after birth
E. clinical cyanosis is usually mild

60. Clinical manifestations of hypoplastic left-heart syndrome include

A. cyanosis always obvious in the 1st 48 hr of life
B. deep-blue color skin
C. bounding peripheral pulses
D. occasional association with Turner syndrome
E. autosomal recessive inheritance is usual

61. Polysplenia (left isomerism) is characterized by
    A. right-sided stomach
    B. decreased pulmonary blood flow
    C. absent gallbladder
    D. howell-jolly bodies
    E. severe cyanosis

62. Pentalogy of Cantrell consists of the following **EXCEPT**
    A. ectopia cordis
    B. midline supra umbilical abdominal defect
    C. deficiency of the anterior diaphragm
    D. defect of the upper sternum
    E. intracardiac defect

63. The **MOST** common cause of pulmonary hypertension in pediatric patients is
    A. idiopathic pulmonary arterial hypertension
    B. pulmonary venoocclusive disease
    C. left-sided valvular heart disease
    D. chronic obstructive pulmonary disease
    E. interstitial lung disease

64. All patients with clinical evidence of a PDA are at increased risk for endocarditis. As result, PDA should be considered for closure **EXCEPT**
    A. PDA with severe irreversible pulmonary hypertension
    B. small, hemodynamically insignificant PDA
    C. small, hemodynamically significant PDA
    D. moderate size PDA
    E. large PDA

65. Phenytoin is used for treatment of
    A. supraventricular tachycardia
    B. atrial fibrillation
    C. atrial flutter
    D. ventricular tachycardia
    E. digitalis intoxication

66. Side effects of propranolol may include
    A. hypothyroidism
    B. elevated triglycerides
    C. school performance problems
    D. hepatic toxicity
    E. pulmonary fibrosis

67. The following criteria are indications for further investigation of premature ventricular contractions (PVCs) that could require suppressive therapy **EXCEPT**

A. multiform PVCs
B. disappear during exercise
C. R-on-T phenomenon
D. extreme frequency of beats
E. presence of underlying heart disease

68. In urgent situations of supraventricular tachycardia (SVT) when symptoms of severe heart failure have already occurred.
Of the following, the initial management is

A. adenosine by rapid intravenous push
B. DC cardioversion (0.5-2 J/kg)
C. verapamil
D. digoxin
E. amiodarone

69. Ventricular tachycardia (VT) is defined as at least 3 premature ventricular contractions PVCs at >120 beats/min. Hemodynamically unstable patients with VT should be immediately treated with

A. catheter ablation
B. ICD implantation
C. DC cardioversion
D. amiodarone
E. procainamide

70. In 90% of cases of endocarditis, the causative agent is recovered from the first 2 blood cultures. Antimicrobial pretreatment of the patient reduces the yield of blood cultures to

A. 10%
B. 30%
C. 40%
D. 60%
E. 80%

71. Despite the use of antibiotic agents, mortality remains high, in the range of 20-25%. Serious morbidity occurs in 50-60% of children with documented infective endocarditis. Of the following, the **MOST** common morbidity is

A. heart failure
B. pulmonary emboli
C. mycotic aneurysms
D. acquired ventricular septal defect

E. heart block

72. Anthracycline cardiotoxicity (doxorubicin [Adriamycin]) on rare occasion causes acute inflammatory myocardial injury, but more classically results in dilated cardiomyopathy (DCM) and occurs in up to 30% of patients given a cumulative dose of doxorubicin exceeding

A. 250 mg/m2
B. 350 mg/m2
C. 450 mg/m2
D. 550 mg/m2
E. 650 mg/m2

73. Restrictive cardiomyopathy (RCM) accounts for <5% of cardiomyopathy cases. RCM is characterized by all the following EXCEPT

A. normal ventricular chamber dimensions
B. normal myocardial wall thickness
C. preserved systolic function
D. normal atrial chamber dimensions
E. high ventricular diastolic pressure

74. Diastolic dysfunction and normal systolic function are recognized features of

A. dilated cardiomyopathy
B. hypertrophic cardiomyopathy
C. left ventricular noncompaction
D. restrictive cardiomyopathy
E. arrhythmogenic right ventricular cardiomyopathy

75. In Asia, the following hepatitis virus appears to be a significant cause of viral myocarditis

A. hepatitis A virus
B. hepatitis B virus
C. hepatitis C virus
D. hepatitis D virus
E. hepatitis E virus

76. The vast majority of tumors originating from the heart are benign.
Of the following, the **MOST** common pediatric cardiac tumors are

A. fibromas
B. rhabdomyomas
C. myxomas
D. hemangiomas
E. papillomas

77. Afterload reducers are not indicated in children with heart failure secondary to
A. cardiomyopathy
B. severe mitral insufficiency
C. aortic insufficiency
D. ventricular septal defect
E. aortic stenosis

78. Adverse reactions to angiotensin-converting enzyme inhibitors (ACEIs) include the following **EXCEPT**
A. hypotension
B. hypokalemia
C. maculopapular pruritic rash
D. renal toxicity
E. chronic cough

79. The definition of hypertension in children is
A. average systolic blood pressure (SBP) and/or diastolic BP that is ≥95th percentile for age, sex, on ≥3 occasions
B. average systolic blood pressure (SBP) and/or diastolic BP that is ≥95th percentile for age, sex, and height on ≥2 occasions
C. average systolic blood pressure (SBP) and/or diastolic BP that is ≥95th percentile for age, sex, and height on ≥3 occasions
D. average systolic blood pressure (SBP) and/or diastolic BP that is ≥90th percentile for age, sex, and height on ≥3 occasions
E. average systolic blood pressure (SBP) and/or diastolic BP that is ≥99th percentile for age, sex, and height on ≥3 occasions

80. Renal childhood diseases responsible for chronic hypertension include
A. hemolytic-uremic syndrome
B. acute tubular necrosis
C. congenital dysplastic kidney
D. pyelonephritis
E. renal trauma

81. Measuring serum potassium is essential in a child with confirmed hypertension because hyperkalemia may be seen in
A. Gordon syndrome
B. Liddle syndrome
C. glucocorticoid remedial aldosteronism
D. apparent mineralcorticoid excess
E. pheochromocytoma

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PART XX The Cardiovascular System ANSWERS

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1.**(C).** CHARGE association (coloboma, heart, atresia choanae, retardation, genital and ear anomalies) is associated with TOF, aortic arch and conotruncal anomalies.
2.**(A).** Marfan syndrome is associated with dilated and dissecting aorta, aortic valve regurgitation, mitral valve prolapsed, William syndrome with supravalvular aortic stenosis, peripheral pulmonary stenosis, Trisomy 21 (Down syndrome) with endocardial cushion defect, VSD, ASD, PDA, while XO (Turner syndrome) associated with coarctation of aorta, aortic stenosis.
3.**(D).** Louder with patient in upright position.
4.**(E).** Common causes are costochondritis, Trauma or muscle overuse/strain, asthma (often exercise-induced), severe cough, pneumonia, reflux esophagitis, anxiety, hyperventilation, precordial catch syndrome (Texidor's twinge), sickle cell vasoocclusive crisis, and idiopathic.
5.**(B).**
6.**(A).** Although dextroposed transposition of the great arteries represents only about 5% of congenital heart defects, it is the most common cyanotic lesion to present in the newborn period.
7.**(A).**
8.**(B).** Hypoplastic left heart syndrome accounts for 1% of all congenital heart defects but is the most common cause of death from cardiac defects in the first month of life.
9.**(E).** Fluid overload, PDA, VSD, and Cor pulmonale (BPD) are causes of heart failure in premature neonate.
10.**(A).** If left-sided failure is predominant, tachypnea, orthopnea, wheezing, and pulmonary edema are seen. Hepatomegaly, edema, and distended neck veins are signs of right-sided failure.
11.**(D).** In otherwise healthy children, neurocardiogenic syncope is most common. This entity goes by a number of terms, including vasovagal syncope, neurally mediated syncope, and autonomic syncope.
12.**(D).** The most common clinical sign of coarctation of the aorta in older children is differential blood pressure: arms > legs (100%).
13.**(B).**
Decreased pulmonary markings (diminished pulmonary blood flow)

- Pulmonary atresia or severe stenosis
- Tetralogy of Fallot
- Tricuspid atresia
- Ebstein anomaly

Increased pulmonary markings (increased pulmonary blood flow)

- Transposition of great arteries
- Total anomalous pulmonary venous return
- Truncus arteriosus

14.**(A).** Endocardial fibroelastosis is a cause after the neonatal period.
15.**(E).** Little change in heart rate with activity, crying, or breath holding.
16.**(C).**
17.**(B).** Dysrhythmias (other than first-degree heart block) is an associated factor.
18.**(E).** Apnea, fever, cutaneous flushing, seizures, hypotension, and bradycardia/tachycardia.
19.**(A).** Gender differences in the occurrence of specific cardiac lesions have been identified. Transposition of the great arteries and left-sided obstructive lesions are slightly more common in boys (≈65%), whereas atrial septal defect, VSD, PDA, and pulmonic stenosis are more common in girls.
20.**(B).** In most patients with an ASD, the characteristic finding is that the 2nd heart sound is widely split and fixed in its splitting during all phases of respiration.
21.**(E).**
22.**(E).** Closure of all supracristal ventricular VSDs at the time of diagnosis is commonly recommended to prevent the development of aortic regurgitation, even in an asymptomatic child.
23.**(C).** The radial and femoral pulses should always be palpated simultaneously for the presence of a radial-femoral delay. Normally, the femoral pulse occurs slightly before the radial pulse. A radial-femoral delay occurs when blood flow to the descending aorta is dependent on collaterals, in which case the femoral pulse is felt after the radial pulse.
24.**(C).** A variety of lesions may give rise to chronic pulmonary venous hypertension, which when extreme may result in pulmonary arterial hypertension and right-sided heart failure. These lesions include congenital mitral stenosis, mitral insufficiency, total anomalous pulmonary venous return with obstruction, left atrial myxomas, cor triatriatum (stenosis of a common pulmonary vein), individual pulmonary vein stenosis, and supravalvular mitral rings.
25.**(E).** This lesion is not progressive in childhood, and specific therapy is not indicated. Antibiotic prophylaxis is no longer recommended during surgery and dental procedures.
26.**(B).** Sinus rate <90 beats/min in neonates and <60 beats/min in older children is considered to be sinus bradycardia.
27.**(E).** Ocular pressure must never be performed, and carotid sinus massage is very rarely effective.
28.**(D).** The other antiprotozoal agent, pentamidine isethionate may cause long Q-T syndrome (LQTS).
29.**(B).** The most common cause of death in competitive athletes is hypertrophic cardiomyopathy, with or without obstruction to left ventricular outflow.
30.**(B).** Viridans-type streptococci (α-hemolytic streptococci) and Staphylococcus aureus remain the leading causative agents for endocarditis in pediatric patients.

31.**(A).** The **Duke criteria** help in the diagnosis of endocarditis. **Major criteria** include (1) positive blood cultures (2 separate cultures for a usual pathogen, 2 or more for less typical pathogens), and (2) evidence of endocarditis on echocardiography (intracardiac mass on a valve or other site, regurgitant flow near a prosthesis, abscess, partial dehiscence of prosthetic valves, or new valve regurgitant flow).
32.**(C).** Lower limit of normal is 70/min, average 125/min, and upper limit is 190/min.
33.**(C).** The presence of diminished pulses in all extremities is associated with pericardial tamponade, left ventricular outflow obstruction, or cardiomyopathy.
34.**(A).** Tall (>2.5 mm), narrow, and spiked P waves are indicative of **right atrial enlargement** and are seen in congenital pulmonary stenosis, Ebstein anomaly of the tricuspid valve, tricuspid atresia, and sometimes cor pulmonale. These abnormal waves are most obvious in leads II, V_3R, and V_1. Similar waves are sometimes seen in thyrotoxicosis. **Broad P waves,** commonly **bifid** and sometimes **biphasic,** are indicative of left atrial enlargement. They are seen in some patients with large left-to-right shunts (ventricular septal defect [VSD], patent ductus arteriosus [PDA]) and with severe mitral stenosis or regurgitation. Flat P waves may be encountered in hyperkalemia.
35.**(D).** One notable exception is the case of severe regurgitant lesions, most commonly of the tricuspid valve. In these lesions (e.g., Ebstein anomaly or severe right ventricular outflow obstruction, the parallel fetal circulation cannot compensate for the volume load imposed on the right side of the heart. In utero heart failure, often with fetal pleural and pericardial effusions and generalized ascites (nonimmune hydrops fetalis) may occur.
36.**(B).** The most common lesions in this group are those that cause left-to-right shunting: atrial septal defect, ventricular septal defect (VSD), AV septal defects (AV canal), and patent ductus arteriosus.
37.**(D).** This group of congenital heart lesions include (tetralogy of Fallot, pulmonary atresia with an intact septum, tricuspid atresia, total anomalous pulmonary venous return with obstruction).
38.**(D).** Arterial oxygen saturation is usually normal unless there is an associated intracardiac shunt.
39.**(B).** Flushing, headache, diarrhea, hypotension, fluid retention, exacerbation of heart failure, anemia, elevated liver function tests, and palpitations are common side effects of bosentan.
40.**(A).**
41.**(B).** Patients who have undergone surgery entailing the use of cardiopulmonary bypass, especially in the newborn period, should be watched carefully during their early school years for signs of mild to moderate learning disabilities, which are often amenable to early remedial intervention. The risk is higher in patients who have undergone repair using hypothermic total circulatory arrest than in those where systemic blood flow is maintained using cardiopulmonary bypass.
42.**(E).** The postpericardiotomy syndrome may occur toward the end of the 1st postoperative week or may sometimes be delayed until weeks or months after surgery.

This febrile illness is characterized by fever, decreased appetite, listlessness, nausea, and vomiting. Chest pain is not always present, so a high index of suspicion should be maintained in any recently postoperative patient.

43.**(E).** Many argue that only for isolated patent ductus arteriosus is total repair really achieved, with no requirement for long-term follow-up.

44.**(A).** Pregnancy is contraindicated in mothers with severe pulmonary hypertension, severe obstructive lesions, and Marfan syndrome, aortic root >40 mm.

45.**(A).** The exception to this rule is patients with small supracristal or perimembranous VSD with associated prolapse of the aortic cusp into the defect resulting in progressive aortic regurgitation. These patients should be considered for surgical repair at the time of diagnosis to prevent progressive aortic valve damage.

46.**(A).** Overall, for those patients who underwent early repair before the development of pulmonary vascular disease, the long-term prognosis is good. The most common long-term complication is left AV valve regurgitation, with approximately 5-10% of patients requiring surgical revision for left AV valve repair or replacement during follow-up.

47.**(E).** A silent PDA is a tiny defect that cannot be heard by auscultation and is only detected by other nonclinical means such as echocardiography. Life expectancy is always normal in this population and the risk for endocarditis is extremely low. Patients with a small PDA have an audible long-ejection or continuous murmur heard best at the left upper sternal border that radiates to the back. In addition, they have normal peripheral pulses. Because there is negligible left to right shunting these patients have normal left aorta (LA) and left ventricle (LV) size and normal pulmonary artery pressure by echocardiography and chest x-ray. These patients like those with silent PDAs are aysmptomatic and live a normal life expectancy. They have a higher risk for endocarditis.

48.**(C).** For those patients with severe stenosis (defined as a peak gradient of >50 mm Hg), the majority ultimately require an intervention, either surgery or balloon valvuloplasty by age 25 yr.

49.**(E).** If cardiac output is significantly decreased, the intensity of the murmur at the right upper sternal border may be minimal.

50.**(E).** In cases that are well defined by echocardiography, CT, or MRI, diagnostic catheterization is not usually required before surgery.

51.**(D).** Congenital mitral stenosis is a rare anomaly that can be isolated or associated with other defects, the most common being subvalvar and valvar aortic stenosis and coarctation of the aorta (Shone complex).

52.**(C).** The spells occur most frequently in the morning on initially awakening or after episodes of vigorous crying. Temporary disappearance or a decrease in intensity of the systolic murmur is usual as flow across the right ventricular outflow tract diminishes. The onset is usually spontaneous and unpredictable. Spells are associated with reduction of an already compromised pulmonary blood flow, which, when prolonged, results in severe systemic hypoxia and metabolic acidosis. Infants who are only mildly

cyanotic at rest are often more prone to the development of hypoxic spells because they have not acquired the homeostatic mechanisms to tolerate rapid lowering of arterial oxygen saturation, such as polycythemia.

53.**(D).** Because metabolic acidosis develops when arterial Po2 is <40 mm Hg, rapid correction (within several minutes) with intravenous administration of sodium bicarbonate is necessary if the spell is unusually severe and the child shows a lack of response to the foregoing therapy.

54 **(C).** Concavity in the region of the main pulmonary artery.

55.**(B).** The severity of symptoms and the degree of cyanosis are highly variable and depend on the extent of displacement of the tricuspid valve and the severity of right ventricular outflow tract obstruction.

56.**(D).** In normally related great vessels, the aorta is posterior and to the right of the pulmonary artery; in d-transposition of the great arteries (d-TGA), the aorta is anterior and to the right of the pulmonary artery (the *d* indicates a dextropositioned aorta, transposition indicates that it arises from the anterior right ventricle).

57.**(A).** Obstructed TAPVR is a pediatric cardiac surgical emergency because prostaglandin therapy is usually not effective.

58.**(A).** In neonates with marked pulmonary venous obstruction, the chest x-ray demonstrates a very dramatic perihilar pattern of pulmonary edema and a small heart.

59.**(D).** When pulmonary vascular resistance is relatively highimmediately after birth, pulmonary blood flow may be normal; as pulmonary resistance drops in the 1st mo of life, blood flow to the lungs is greatly increased and heart failure ensues.

60.**(D).** Although cyanosis may not always be obvious in the 1st 48 hr of life, a grayish-blue color of the skin is soon apparent and denotes a mix of cyanosis and poor perfusion. All of the peripheral pulses may be weak or absent. This lesion may be isolated or associated in 5-15% of patients with known genetic syndromes, such as Turner syndrome, trisomy 13, 18, or 21, Jacobsen syndrome (11q deletion), Holt-Oram syndrome, andRubinstein-Taybi syndrome. Occasionally it is familial and inherited as an autosomal recessive trait.

61.**(C).** All other options are features of asplenia (right isomerism).

62.**(D).** Defect of the lower sternum.

63.**(A).** Idiopathic or familial disease is the most common in pediatric patients (~55%), followed by pulmonary hypertension secondary to congenital heart disease (~35%) and chronic respiratory disorders (~15%).

64.**(A).** All PDAs except for small silent PDAs and those patients with severe irreversible pulmonary hypertension should be considered for closure.

65.**(E).** Other options are treated by procainamide.

66.**(C).** Other options are side effects of amiodarone.

67.**(B).** It is important to distinguish PVCs that are benign from those that are likely to lead to more severe arrhythmias. The former usually disappear during the tachycardia of exercise. If they persist or become more frequent during exercise, the arrhythmia may have greater significance.

68.**(B).** In urgent situations when symptoms of severe heart failure have already occurred, synchronized DC cardioversion (0.5-2 J/kg) is recommended as the initial management.
69.**(C).**
70.**(D).** Antimicrobial pretreatment of the patient reduces the yield of blood cultures to 50-60%. The microbiology laboratoryshould be notified if the patient has received antibiotics so thatmore sophisticated methods can be used to recover the offending agent.
71.**(A).** The most common is heart failure caused by vegetations involving the aortic or mitral valve. Myocardial abscesses and toxic myocarditis may also lead to heart failure without characteristic changes in auscultatory findings and, occasionally, to life-threatening arrhythmias.
72.**(D).** Exceeding 550 mg/m2. The risk of toxicity appears to be exacerbated by concomitant radiation therapy.
73.**(D).** Dramatic atrial dilation can occur as a result of the abnormal myocardial compliance and high ventricular diastolic pressure.
74.**(D).**
75.**(C).** Coxsackievirus and other enteroviruses, adenovirus, parvovirus, Epstein-Barr virus, parechovirus, influenza virus, and cytomegalovirus are the most common causative agents in children, though most known viral agents have been reported. In Asia, hepatitis C virus appears to be significant as well.
76.**(B).** Rhabdomyomas are the most common pediatric cardiac tumors and are associated with tuberous sclerosis in 70-95% of cases.
77.**(E).** ACEIs and ARBs are not generally used in the presence of stenotic lesions of the left ventricular outflow tract because of concern over coronary perfusion.
78.**(B).** Adverse reactions to ACEIs include hypotension and its sequelae (weakness, dizziness, syncope) and hyperkalemia.
79.**(C).** The National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents published the Fourth Report on the Diagnosis, Evaluation, and Treatment of High Blood Pressure in Children and Adolescents (Fourth Report) in 2004.
The Fourth Report defined hypertension as average systolic blood pressure (SBP) and/or diastolic BP that is ≥95th percentile for age, sex, and height on ≥3 occasions. Prehypertension was defined as average SBP or diastolic BP that are ≥90th percentile but <95th percentile.
80.**(C).** Other options are responsible for acute/ intermittent hypertension.
81.**(A).** Measuring serum potassium is essential because hypokalemia may be present in Liddle syndrome, glucocorticoidremedial aldosteronism, and apparent mineralcorticoid excesssyndrome, while hyperkalemia may be seen in Gordon syndrome.